Pure Arterial Malformation of the Left Middle Cerebral Artery: A Case Report

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Case Report

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Abstract

Background: Pure arterial malformations (PAMs) are rare intracranial vascular anomalies and involvement of the distal M3 segment of the middle cerebral artery are also uncommon. Diagnosis can become elusive when PAMs occur in atypical locations due to insufficient knowledge of the disease in spite of specific imaging characteristics. PAMs actually have a benign natural history and should be managed conservatively. We present a patient with a PAM of the left middle cerebral artery that was mistaken for an arteriovenous malformation and embolized.

Case presentation: We report a case of pure arterial malformation of the left middle cerebral artery in a 41-year-old man with a two-day history of dizziness and limb convulsions. Preoperative MRI and DSA displayed the left middle cerebral artery was dilated, overlapping, and tortuous arteries forming a mass of arterial loops with a coil-like appearance. He was mistaken for an arteriovenous malformation and embolized.

Conclusion: Our case highlights that intracranial vascular anomalies forming a mass of arterial loops lacking a venous component should be considered pure arterial malformations. Management and individual therapy are administrated to patient who are screened out PAMs.

Background

Pure arterial malformations (PAMs) are rare intracranial vascular anomalies composed of dilated, overlapping, and tortuous arteries which form a mass of arterial loops. They lack a venous component and have a coil-like appearance on imaging studies. PAMs actually have a benign natural history and should be managed conservatively. We present a patient with a PAM of the left middle cerebral artery that was mistaken for an arteriovenous malformation and embolized.

Case Presentation

A 41-year-old previously healthy man who presented to an outside facility with a two-day history of dizziness and limb convulsions was transferred to our hospital for further management. Computed tomography (CT) of the head showed a tubular-like slightly high-density lesion and parenchymal calcifications in the left occipital lobe; sulcal spaces in the left hemisphere were prominent (Fig. 1). Axial T1-weighted imaging revealed hypointensity in the left occipital lobe with a small nodular area of hyperintensity (Fig. 2a). Axial T2-weighted imaging demonstrated dilation of the left middle cerebral artery and a vascular malformation in the occipital lobe (Fig. 2b). Magnetic susceptibility-weighted imaging(SWI) showed patchy areas of low signal intensity within the lesion and a dilated tortuous left middle cerebral artery (Fig. 2c). Three-dimensional time-of-flight magnetic resonance angiography(MRA) showed moderate dilation of the left M1 segment with an aneurysm (Fig. 2d). Post-contrast T1-weighted imaging showed multiple enhancing vessels within the lesion that were in continuity with the thickened left middle cerebral artery (Fig. 2e). Three-dimensional post-contrast isovoxel reconstruction showed left

middle cerebral artery dilation with one ring and one aneurysm (Fig. 2f). Diffusion tensor imaging showed sparse and decreased number of white matter fiber tracts in the left temporoparieto-occipital area (Fig. 2g).Digital subtraction angiography (DSA) showed a dilated, corkscrew-like left middle cerebral artery with distal tortuous vessels and coil-like structure (Fig. 3). Arteriovenous malformation was diagnosed and endovascular embolization was performed using liquid glue. Two days after the procedure, computed tomography showed multiple areas of high signal density within the left temporoparieto-occipital lobe (Fig. 4). The patient h was discharged on sodium valproate for seizure prophylaxis. Since the procedure, the patient has had no recurrence of symptoms.

Discussion

The patient presented in this case report had a PAM that was misdiagnosed as an arteriovenous malformation. PAM is a rare vascular malformation, which is defined as dilated, overlapping, and tortuous arteries forming a mass of arterial loops with a coil-like appearance in the absenceof arteriovenous shunting[1–3]. PAMs are rare and usually occur in young women. They frequently involve the distal branches of the anterior or posterior cerebral artery[1, 3–5]. Involvement of the distal M3 segment of the middle cerebral artery, as in our patient, is rare.

Previous studies have indicated that PAMs of specific cerebral arteries frequently have certain characteristics. For example, PAMs of the middle and posterior cerebral artery and the posterior communicating artery often have a tight vascular circle with multiple aneurysms and calcification. Those of the distal anterior cerebral artery tend to be moderately curved with less dense coils and often contain calcification. Superior cerebellar and posterior inferior cerebellar artery PAMs often lack substantial dilation and are accompanied by aneurysms[6].The PAM in our patient involved the distal left middle cerebral artery.

As blood flows through the spiral collection of arteries in a PAM, it slows down and gradually rotates, causing arterial dilation and clot formation. Arterial elongation and angulation can cause stretching and distortion of arterial branch openings, which can result in reduced blood flow to the perfused region and consequent cerebral infarction and atrophy. Our patient had evidence of cerebral infarction in the involved brain area and a small amount of hemorrhage.

Although the cause of PAMs remains unclear, several hypotheses have been proposed. Congenital factors such as an inherited defect or insult during brain development can result in arterial dysplasia. Acquired factors such as somatic mutations occurring later in life, viral infection, chronic healed dissection, and inflammatory, immune-related, and degenerative factors may also play a role[5, 6].

PAMs are often found incidentally. However, our patient was symptomatic with dizziness and convulsions, which may have been caused by left temporoparietal hemosiderin deposition and fiber bundle damage[7].

PAMs are often misdiagnosed as arterial prolongation or dilation or arteriovenous malformations. Arterial prolongation and dilation are more common in elderly men with a history of hypertension and smoking. Although the involved blood vessels are dilated and curved in patients with arterial prolongation and dilation, vessels of the vertebrobasilar circulation vessels and internal carotid artery are usually affected. PAM vessels are severely curved and overlapped, which leads to a large number of arterial loops [8]. Draining veins are seen in arteriovenous malformations [9].

Previous studies have demonstrated that PAMs do not usually progress; therefore, conservative treatment of incidental PAMs is recommended [5]. However, those associated with an aneurysm should be closely monitored, as aneurysm treatment may be necessary if imaging progression is observed [10].

Conclusion

In this case, we have demonstrated the imaging characteristics of PAMs in the intracranial. Management and individual therapy are administrated to patient who are screened out PAMs.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

The patient has given consent for the case report.

Availability of data and materials

All data and material are included in this published article.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

Nian-xia Fu and Jian-xun Song wrote the main manuscript text. Jian-xun Song Ying-te Wu and Guo-hui Lin prepared figures 1-4. All authors reviewed and approved the final manuscript.

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Figures

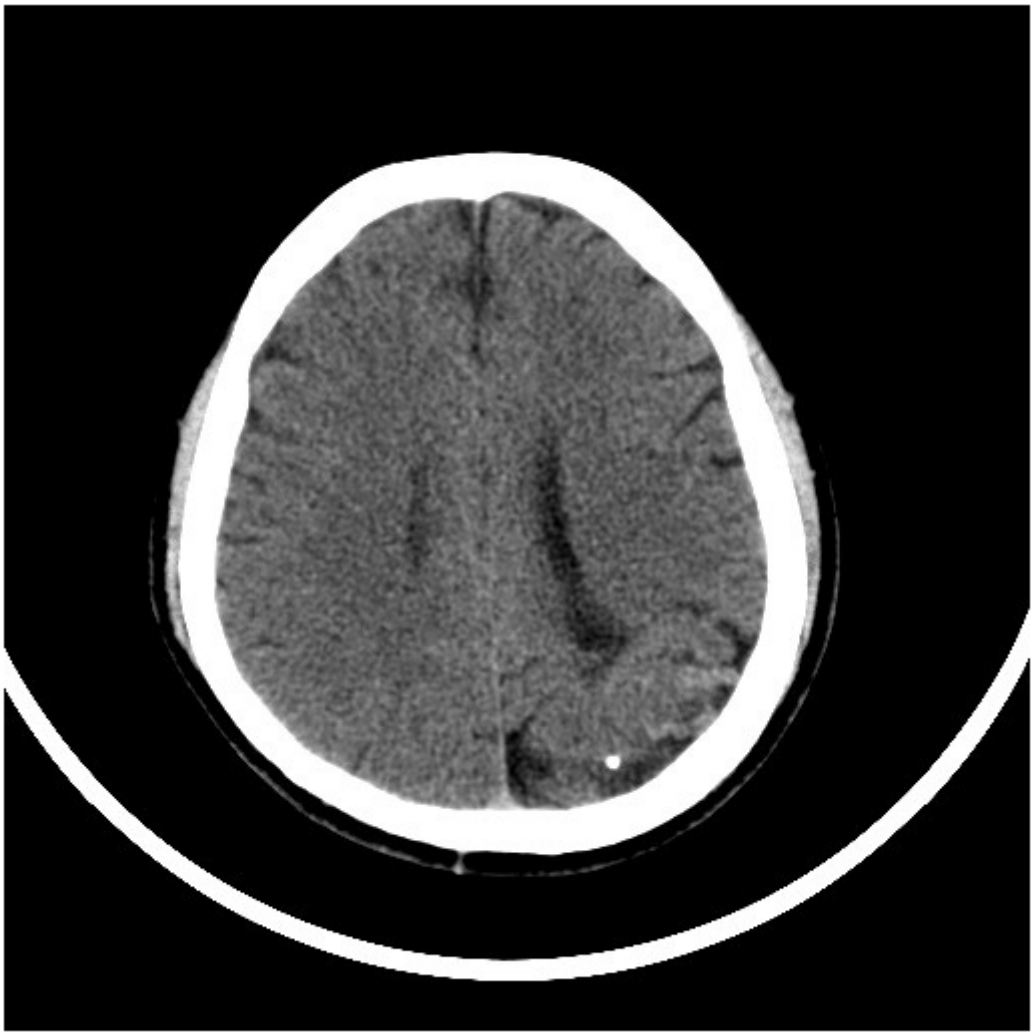


Figure 1

Axial head computed tomography. Strips and nodules of slightly high density with scattered punctate calcifications can be seen in the left parieto-occipital lobe.

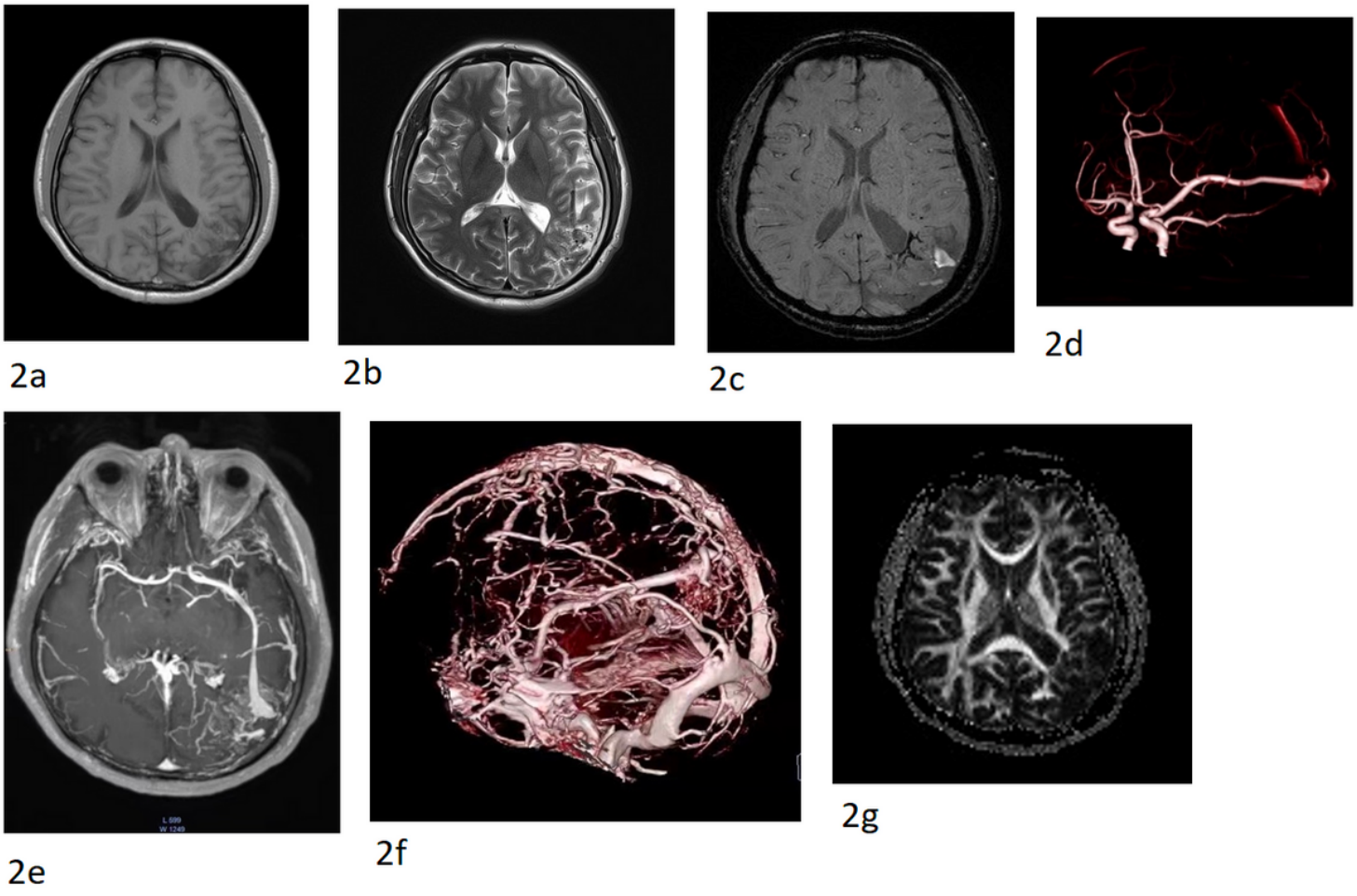


Figure 2

2a. Axial T1-weighted imaging. The left parieto-occipital lobe shows low signal intensity with a small nodular area of high signal intensity.

2b. Axial T2-weighted imaging. The left temporoparietal lobe shows a worm-like blood vessel flow void. The left middle cerebral artery flow void is also visualized.

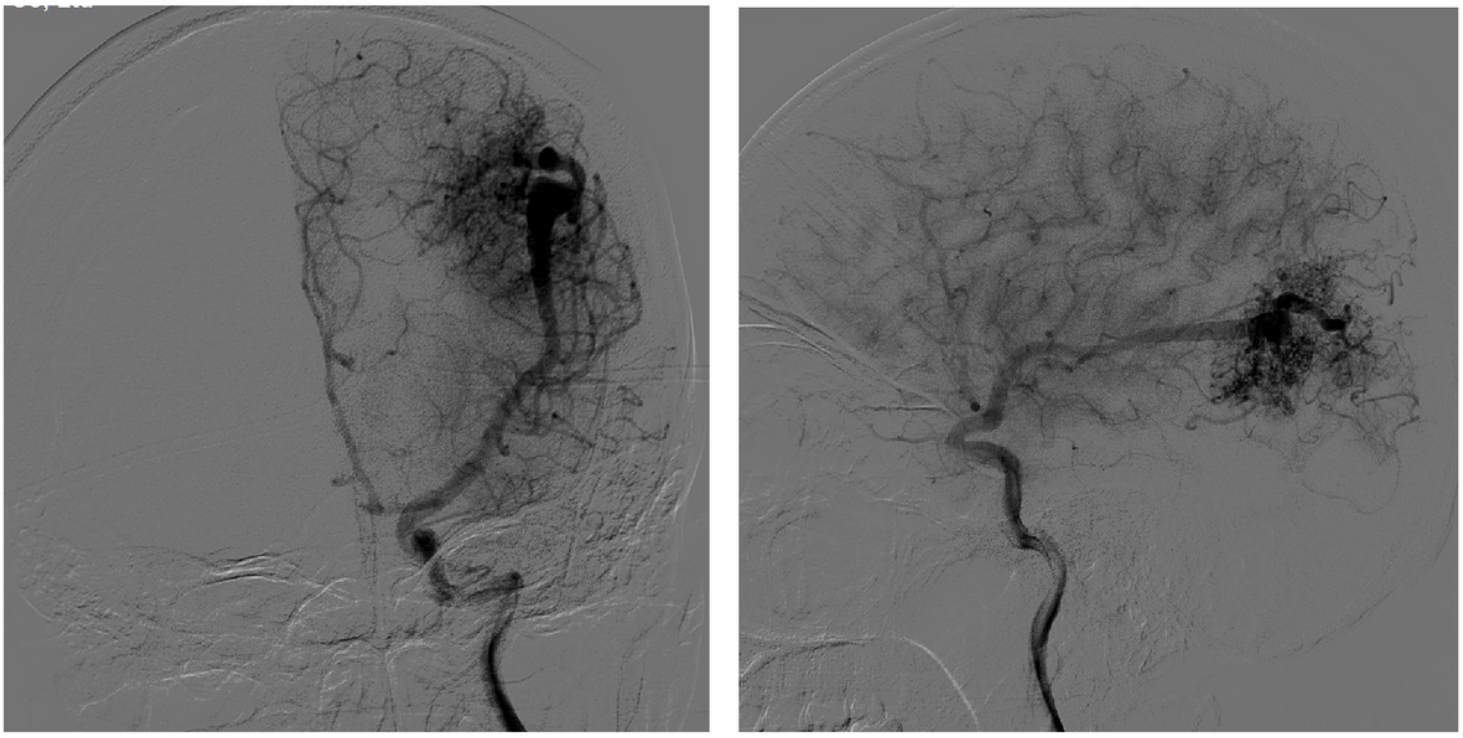
2c. Fig. 2h. Axial SWI. A patchy low signal intensity within the lesion and dilation of the tortuous left middle cerebral artery are seen.

2d. 3D TOF MRA shows widening and distal dilation of the left middle cerebral artery.

2e. Axial post-contrast T1-weighted imaging. Multiple enhancing vascular structures in continuity with the thickened left middle cerebral artery are seen in the left temporoparieto-occipital lobe.

2f. Three-dimensional post-contrast isovoxel reconstruction. The left middle cerebral artery appears dilated with a ring and an aneurysm.

2g. Diffusion tensor imaging. The white matter fiber bundles in the left temporoparieto-occipital lobe appear sparse and reduced in number.



3a 3b

Figure 3

DSA with anteroposterior (a) and lateral (b) views after left internal carotid artery injection. The left middle cerebral artery is dilated proximal to a coiled mass of dilated, elongated, and tortuous vessels. No early venous drainage is seen, suggestive of a pure arterial malformation.

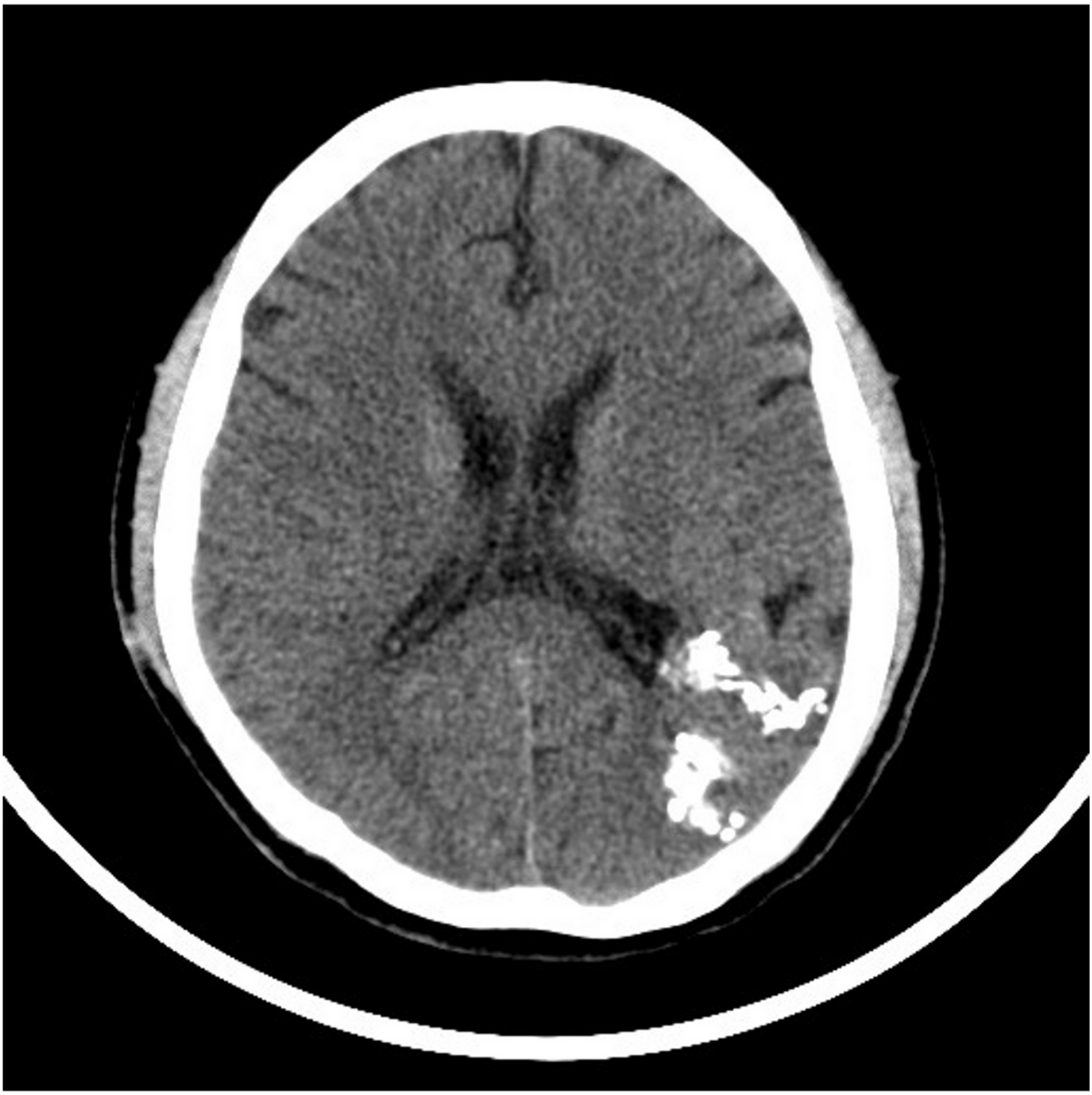


Figure 4

Axial computed tomography after embolization. Multiple patchy areas of high-density are seen in the left temporoparieto-occipital lobe.